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Large pseudoencapsulated subcutaneous angiomyxoma: Surgical management.

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Superficial angiomyxoma is a rare benign skin tumor of mesenchymal origin composed of a myxoid matrix, thin-walled blood vessels, and sparse small spindle cells. This tumor is reported to have a high local recurrence rate when excised, possibly because muscle and soft tissue infiltration can occur. Thus, the term *superficial* can be misleading. In one published series, the local recurrence rate was 36%[1] and in another 38%.[2] However, all reported series are pathologic studies in which details of surgical excisions are scant. We describe a large seemingly encapsulated angiomyxoma that we removed in toto sparing overlying skin, after which large peripheral Mohs-type horizontal frozen sections were removed in selected areas of surrounding soft tissue to help confirm complete excision.

Abbreviations used:

MMS: Mohs micrographic surgery
Rb: retinoblastoma

CASE REPORT

A 68-year-old man without any significant medical problems presented with a right chin mass (Fig 1, *A*). The lesion was asymptomatic and had been present for 10 weeks, during which time it grew rapidly. At the time of physical examination, the mass measured 2 × 5 cm. It was soft, mobile, and appeared to be subcutaneous. There were no changes in the overlying skin and no palpable connection with intraoral structures. A recent ultrasound scan showed a well-defined moderately dense mass above the mandibular margin that was not invading or compressing deeper structures. Color Doppler showed mostly peripheral vascularity.

At the time of incisional biopsy, the tumor appeared as a gelatinous gray mass within subcutaneous tissue without a capsule (Fig 1, *B*). Frozen section and permanent section histopathology of the biopsy specimen stained with hematoxylin and eosin showed small, elongated blood vessels and scattered spindle cells within a myxoid stroma consistent with angiomyxoma (Fig 2, *A*). The spindle cells showed neither atypia nor mitoses. The myxoid stroma also stained metachromatically with toluidine blue on frozen sections. Permanent section immunostaining for retinoblastoma (Rb) nuclear antigen displayed positivity for Rb antigen in 25% to 50% of the spindle cells (Fig 2, *B*).

Once the diagnosis was confirmed on permanent sections as an angiomyxoma, we performed an excision under local anesthesia. To preserve as much normal skin as possible, we made a vertical incision overlying the lesion in the maximum skin tension lines. The tumor appeared to be encapsulated. Thus, we did not think it necessary to needlessly sacrifice the overlying skin. The angiomyxoma, with what appeared to be an intact capsule, was carefully dissected out completely in one piece as a spheroid. There was no abnormal remaining tissue that was seen or felt in the wound cavity. The excised tumor measured 2.3 cm × 2.6 cm × 3.0 cm (Fig 3, *A* and *B*) and was sent for permanent sections. Because of reports in

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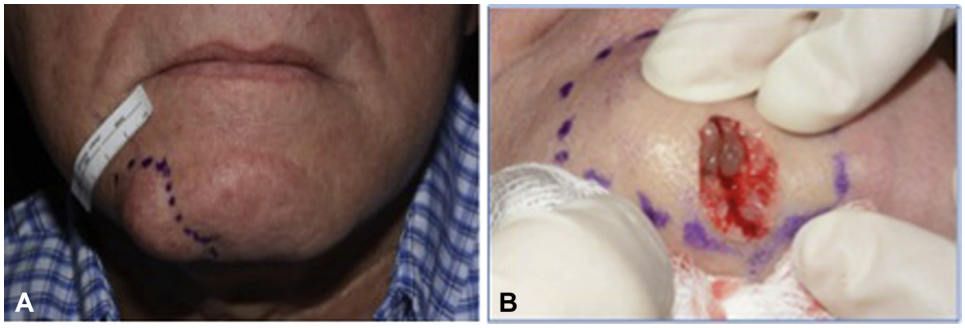

Fig 1. **A**, Poorly demarcated 2- x 5-cm soft mobile tumor on right chin. **B**, Gelatinous gray tissue within subcutaneous layer at the time of incisional biopsy.

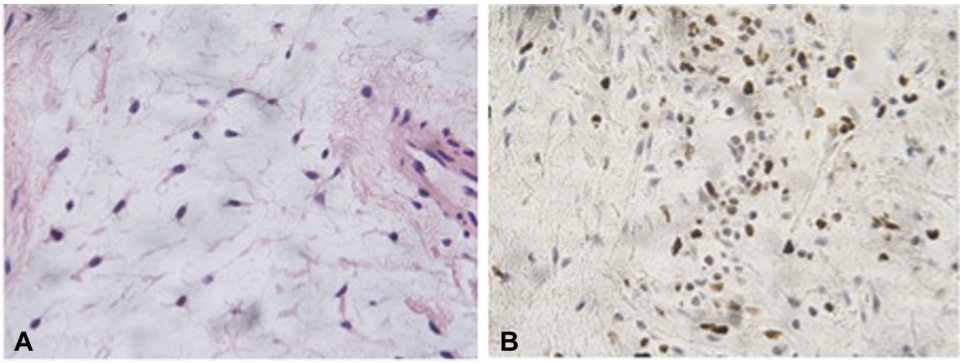

Fig 2. **A**, Sparse spindle cells without atypia within abundant myxoid stroma. One thin-walled blood vessel is seen on right. **B**, Rb nuclear antigen immunostaining positive in about 40% of spindle cells. (**A** and **B**, Hematoxylin-eosin stain; original magnifications: ×40.)

the literature that angiomyxoma infiltrates subcutaneous tissue and muscle, we thought it prudent that additional tissue be excised in selected areas and checked for infiltration. More specifically, this excision included 2 large but thin horizontal layers similar to Mohs micrographic surgery layers, that were taken within the wound cavity beneath the overlying skin and on top of the underlying muscle. Five additional small tissue samples of the wound cavity were also excised. The 2 Mohs-type specimens were flattened and subdivided into 5 frozen sections, similar to flattened Mohs en face frozen sections, color-coded, and stained with toluidine blue, which highlights the myxoid stroma. No myxoid-type material was seen either on the 5 Mohs-type sections or the 5 additional small tissue samples. The frozen section blocks were not sent for permanent sections. The wound was then sutured (Fig 4). The wound healed well, and there has been no recurrence after 6 months.

On permanent hematoxylin-eosin–stained sections, a cross-section of the encapsulated angiomyxoma showed it was bounded on 2 sides by muscle fascia but extended to the cut margins on the other 2 sides. However, the Mohs-type frozen sections beyond these 2 sides did not contain myxoid material. Further, because the muscle fascia was the clinical capsule, the capsule was really a pseudocapsule.

DISCUSSION

Angiomyxomas are commonly found in middle-aged men and are usually slow-growing asymptomatic lesions.[2,3] Clinically, these growths can appear similar to a variety of soft tissue tumors; thus, tissue biopsy for histopathologic diagnosis is required to make the diagnosis.[2,3] Histopathology findings show a myxomatous matrix in the dermis or subcutaneous tissue, within which are spindled or stellate cells without significant cytologic atypia along with numerous thin-walled blood vessels. Immunohistochemistry often displays variable positivity for S-100, CD34, smooth muscle actin, desmin, and keratin markers.[2,3]

Rb nuclear antigen immunostaining can help to differentiate spindle cells in angiomyxoma from the similar-appearing spindle cells in spindle cell lipoma, which displays total loss of Rb antigen.[4] In our case, depending on the microscopic field, 25% to 50% of the spindle cells were positive for Rb antigen

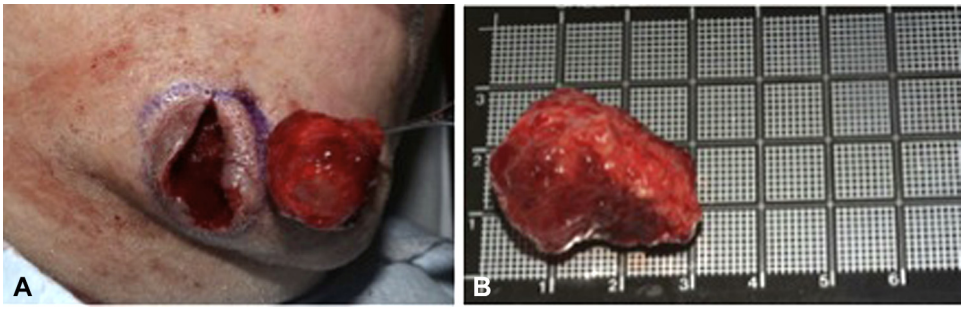

Fig 3. **A**, Excised spheroidal specimen dissected free of overlying skin and surrounding tissue as one piece that appears to be encapsulated. **B**, Excised spheroidal specimen measured 2.3 cm × 2.6 cm × 3.0 cm.

Fig 4. Sutured wound with Penrose drain.

immunostaining, thus helping to confirm the diagnosis of angiomyxoma. Other histologic differential diagnoses include aggressive angiomyxoma, soft tissue myxoma, angiomyolipoma, myxoid nerve sheath tumor, myxoid neurofibromas, and angiomyxofibrosarcoma.

Although angiomyxomas are classified as benign, they have a high recurrence rate after excision: 36% in one study[1] and 38% in another study.[2] Sometimes lesions may infiltrate muscle.[1] For this reason, Mohs micrographic surgery (MMS) was recently proposed for obtaining definitively negative peripheral and deep excision margins.[3,5] Although this surgical approach may be feasible for a small superficial angiomyxoma involving the overlying skin as shown in one report,[5] our case points out that large and deep facial lesions and those that appear to be well encapsulated and not attached to overlying skin may not be best suited for MMS. MMS used here would have sacrificed needlessly a large amount of uninvolved overlying skin, underlying muscle, and nerve, thus causing a significant cosmetic and potentially functional deformity. Rather, careful blunt dissection of the lesion without rupture followed by extensive Mohs-type frozen sections allowed for adequate but not complete margin control. Furthermore, when evaluating the article by Calonje et al[1] who evaluated 39 cases of angiomyxoma that were treated by conventional excision, it appears that the mean size (3.20 cm) of the 7 lesions that recurred was larger than the mean size (2.05 cm) of the nonrecurrent lesions. Because our pathologic specimen was 3 cm, it was in the high-risk category for recurrence. Thus, although MMS can certainly be considered for the removal of small angiomyxomas involving the overlying skin, in other instances in which the lesions are large, subcutaneous, and seemingly encapsulated, careful blunt dissection followed by horizontal en face Mohs-type frozen sections might be preferable to preserve normal tissue and provide a good cosmetic result.

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